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CASE REPORTS

ABERNETHY MALFORMATION TYPE II IN A 70-YEAR-OLD PATIENT WITH ANGINA PECTORIS

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Abstract. *Congenital extrahepatic portosystemic shunts (ECPSS) are rare developmental anomalies in which a variable portion of the portal blood bypasses the liver and is shunted in the systemic circulation via one or more aberrant vessels. We present a clinical case of a 70-year-old man, who was referred to the Cardiology Department because of exertional dyspnea, fatigue, and feeling of heaviness and pressure behind the sternum. MDCT of the aorta was performed and an aberrant vessel was discovered with communication with the left iliac vein on one side and superior mesenteric and splenic veins on the other. The portal vein was hypoplastic. The radiologic findings were suggestive of malformation of Abernethy. The ECPSS can be classified into 2 main groups (with complete and partial shunting). The patients have different clinical presentation. Some of them are completely asymptomatic while in others the shunt can manifest even before birth as fetal growth retardation or in the early neonatal period with neonatal cholestasis and galactosemia. Common complications are hepatic encephalopathy and hepatopulmonary syndrome and there is a wide variety of concomitant abnormalities. The imaging modalities play a crucial role in the diagnosis, classification, follow-up and the proper choice of therapeutic management in patients with ECPSS.*

Key words: *Abernethy malformation, portal vein, shunt, congenital*

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INTRODUCTION

Congenital extrahepatic portosystemic shunts are rare developmental anomaly first described by John Abernethy in 1793 [1]. In this anomaly, a variable portion of the portal blood bypasses the liver and is shunted in the systemic circulation via one or more aberrant vessels [2].

CLINICAL CASE

A 70-year-old man was referred to the Cardiology Department because of exertional dyspnea, fatigue, and feeling of heaviness and pressure behind the sternum. His past medical history included benign prostatic hyperplasia and long-lasting arterial hypertension. The blood pressure control was suboptimal, and the patient had repeatedly hypertonic crisis.

At admission, the patient was in good general condition, eupneic, with a regular heart rate and normal pulmonary and heart auscultation. There were no changes upon examination of the abdomen. The extremities were warm with normal arterial pulsations and without swelling.

The electrocardiography revealed sinus rhythm with a heart rate of 80/min, horizontal electrical position, and pathological Q-wave with ST-elevation up to 1 mm in III and aVF-leads.

The complete blood count, blood chemistry, electrolytes, and coagulation profile were normal. Troponin I was negative.

A diagnostic percutaneous selective coronary angiography was performed, which revealed mild atherosclerotic changes without significant stenosis.

The chest radiograph showed clear lungs and pleural spaces with a slightly prominent shadow of the ascending aorta.

Initial workup with echocardiography reported moderate left ventricular hypertrophy with altered segmental kinetics with preserved global systolic function. Because of the ectasia of the ascending aorta and suspicion for intimal flap distal to the left subclavian artery, CT aortography was performed.

The native CT scanning did not reveal hyperdense areas in the aortic wall. Therefore, an intramural hematoma was excluded. The aorta was homogeneously opacified without filling defects and intimal flap. The maximum diameters of the ascending aorta were 42/43 mm. The descending thoracic and abdominal aorta had diameter within reference values. The coronary, visceral, renal, and iliac arteries were homogeneously opacified without stenosis.

During the detailed examination of the abdominal cavity, an aberrant vessel was observed with communication with the left iliac vein on one side and superior mesenteric and splenic veins on the other (Fig. 1).

The portal vein was hypoplastic – its axial diameters were up to 5 mm. The hepatic artery had normal morphology and course. The liver had a normal size, without focal or infiltrative lesions. There were no features for hepatic cirrhosis and ascites (Fig. 2). The remaining abdominal organs did not show any pathological findings.

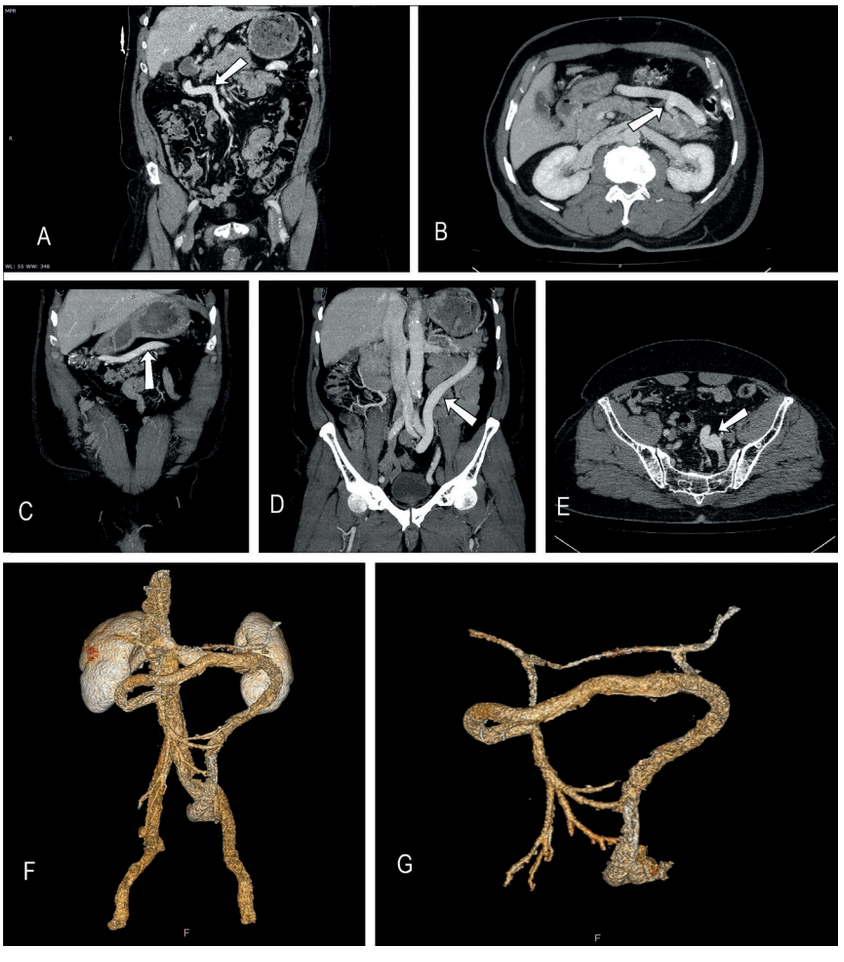

Fig. 1. Contrast enhanced CT scan of the abdomen shows a portosystemic shunt. **A** – the communication between the superior mesenteric vein and the aberrant vessel (arrow). **B** – the drainage of the splenic vein in the shunt across the tail of the pancreas (arrow). **C** – the initial course of the aberrant vessel in the abdominal cavity is from right to left parallel to the transverse colon (arrow). **D** – coronal image shows that the aberrant vessel descends toward the left flank. **E** – axial image shows the communication site between the shunt and the left iliac vein (arrow). **F** – 3D Volume Rendering reconstruction of the venous system of the abdomen. **G** – 3D VR image shows the hypoplastic portal vein and the aberrant portosystemic shunt

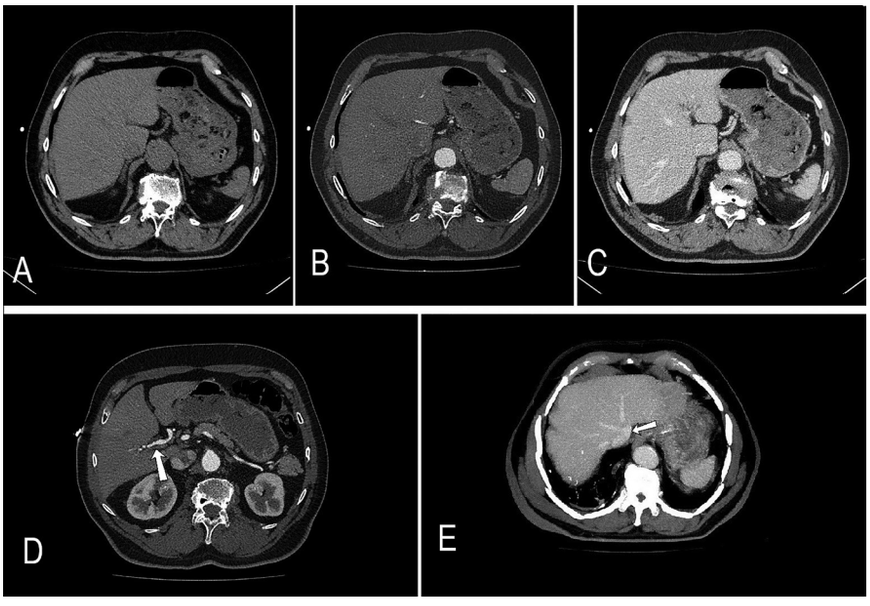

Fig. 2. Contrast enhanced CT scan of the abdomen shows the liver and the hepatic hilum. Native scanning (**A**), arterial phase (**B**), and portal venous phase (*C*) of the liver do not show any focal or infiltrative changes. **D** – axial image from contrast enhanced CT of the liver in arterial phase – the portal vein (arrow) is behind the hepatic artery and the common bile duct in the hilum of the liver. **E** – the inferior vena cava (arrow) and the hepatic veins are normal

DISCUSSION

The radiologic findings were suggestive of malformation of Abernethy. The patient did not have clinical and laboratory findings indicative of possible cardiovascular and neurological complications. Consequently, after a multidisciplinary meeting and thorough discussion of the patient's clinical and radiologic findings, a decision for regular follow-up was made. A strict control of the arterial hypertension is mandatory and antianginal therapy was prescribed.

The normal venous drainage from the abdominal cavity consists of two separate circulations without anatomic communication. The portal vein accounts for 75% of the blood flow to the liver. It is formed by the joining of the splenic and superior mesenteric veins and accepts blood from the inferior mesenteric, gastric, and other veins. It drains blood to the liver from the gastrointestinal tract, except for the rectum, the gallbladder, pancreas, and spleen [1, 2].

In Abernethy malformation a variable portion of the intestinal blood and the carried toxins bypasses the liver and drains into the systemic circulation via one or several aberrant vessels leading to different clinical manifestations and complications [3]. The true incidence is not known for certain – it was estimated to be 1:30 000 births and 1:50 000 permanent shunts [3, 4].

Portosystemic shunts are congenital or acquired vessel communications between the portal and systemic venous system. They can be classified into two main groups – intra- and extrahepatic. The intrahepatic portosystemic shunts have uncertain etiology and are more often found in the right hepatic lobe and can be either congenital or acquired due to trauma or portal hypertension [1].

The extrahepatic congenital portosystemic shunts (ECPSS) are rare and have different morphology and clinical course. They could be classified into 2 main groups with several subdivisions:

- Type I – complete shunting with an absence of intrahepatic portal venous flow;
- Ia – separate drainage of the superior mesenteric and the splenic vein in systemic veins;
- Ib – the superior mesenteric and the splenic veins form short extrahepatic portal vein which drains in a systemic one;
- Type II – incomplete shunting with partially preserved intrahepatic portal venous flow [1, 3].

The most common cause for acquired portosystemic shunts is portal hypertension. There are different routes for portosystemic shunts (gastroesophageal, paraesophageal, paraumbilical, etc.) [1].

There is a wide variety of clinical symptoms and complications. Some patients are completely asymptomatic, and the shunt is found during an imaging test for another reason. In other cases, the shunt can manifest even before birth as fetal growth retardation [3].

The patients can have galactosemia, hyperbilirubinemy, or hyperammonemia due to decreased hepatic metabolism of these substances [5]. The increased levels of nitrogen products can cause hepatic encephalopathy, characterized by tremor, extrapyramidal symptoms, and altered sensorium [2, 5]. In

children, the encephalopathy can demonstrate as lethargy or confusion, seizures, abnormal behavior, mental retardation, etc. The symptoms, however, can be transient [6].

Other patients have symptoms of hepatopulmonary syndrome and portopulmonary hypertension [1, 3]. Hepatopulmonary syndrome is characterized by the triad – arterial deoxygenation, vascular dilation in the lungs, and hepatic disease. It occurs due to the vasoactive substances which bypass the liver and are shunted to the systemic circulation. The patients have dyspnea, cyanosis, and digital clubbing [2]. Portopulmonary hypertension appears later in the course of hepatopulmonary syndrome, and has a poor prognosis [3].

Other patients may present endocrine abnormalities due to decreased hepatic metabolism [16]. They can have hyperandrogenism - primary amenorrhea, early pubarche, and symptoms of virilisation [2, 7]. The reduced insulin clearance can lead to hypoglycemia, and the decreased production of thyroxin-binding globulin can result in hypothyroidism [7].

Due to the absence or decrease in the portal venous blood flow, the arterial blood flow increases. The altered hemodynamics can lead to focal nodular hyperplasia, which is one of the most common lesions in patients with malformation of Abernethy. Others are nodular regenerative hyperplasia, hepatic adenomas, hepatoblastoma, hepatocellular carcinoma, and hepatic cirrhosis [8, 9].

When the shunt ends in iliac veins, gastrointestinal bleeding is more common due to colonic and rectal varices. A rare complication is the membranoproliferative glomerulonephritis, which is due to a decreased clearance of the immune complexes in the liver [3].

The correlation between congenital or acquired heart diseases and CPSS determines the possible presentation with symptoms from the cardiovascular system in the first months of life [6, 7].

The early diagnosis of this malformation is extremely important to prevent some complications. Doppler ultrasound has the advantage of a safe and noninvasive method for assessment of the intrahepatic vessels and can visualize the hemodynamics in the aberrant vessels [2].

CT and MRT can classify the malformation of Abernethy and find concomitant anomalies [1]. MDCT has excellent spatial resolution and allows the visualization of the normal anatomy of the portal vein and its variants. It can show the presence or absence of portal vein and its diameter as well as the communication between the portal and systemic circulation, which can occur at different levels – inferior vena cava, renal and iliac veins, and even the coronary sinus [2]. Main disadvantages of CT are the use of ionizing radiation and the application of contrast material, which can cause allergic reactions, and is potentially nephrotoxic [1].

MRT has its advantages – it does not use ionizing radiation, and allows proper characterization of the hepatic lesions. The examination, however, is longer and with lower spatial resolution [1, 2]. Brain MRI can demonstrate signal alteration in the basal ganglia due to portal encephalopathy [6].

Angiography can detect the small portal vein and its branches, but it is usually performed before endovascular treatment [8]. Occlusion test is also used to evaluate the portal pressure during occlusion of the shunt. This test can help to differentiate between type I and type II extrahepatic portosystemic shunts. Per-rectal scintigraphy use the shunt index to quantify the shunt [10]. The liver biopsy can demonstrate small portal vessels not seen by the imaging modalities [8].

The congenital extrahepatic portosystemic shunts are associated with some cardiovascular, gastrointestinal, skeletal, and genitourinary anomalies [3, 9] (Table 1).

Table 1. Common concomitant anomalies in patients with congenital extrahepatic portosystemic shunts

Cardiovascular abnormalities	Atrial septal defect Ventricular septal defect Patent foramen ovale Coarctation of the aorta Tetralogy of Fallot Patent ductus arteriosus Splenic artery aneurysms Coronary artery fistulas Primitive hypoglossal artery
Gastrointestinal abnormalities	Biliary atresia Choledochal cyst Annular pancreas Duodenal atresia Juvenile polyposis
Urogenital anomalies	Renal agenesis Multicystic dysplastic kidney Bilateral ureteropelvic stenosis Vesicoureteral reflux
Genetic syndromes	Down Turner Leopard Rendu-Osler-Weber
Miscellaneous	Polysplenia Heterotaxy

The method of choice for treatment of type I ECPSS is hepatic transplantation [2]. For type II there are different possibilities – conservative management, transcatheter embolization, and surgical treatment (ligation of the shunt) [1, 3]. The closure of the shunt is mandatory when there are complications and can prevent the development of hepatic encephalopathy and restore the normal portal venous flow [6]. For intrahepatic shunts, when the endovascular treatment is not possible, liver resection is an option [3]. Conservative management includes protein restriction, lactulose, and clinical and imaging follow-up [7].

Disclosure summary

The authors have nothing to disclose.

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